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## Slow-Growing Solitary Bulla of the Upper Arm

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### Clinical Case

A 13 year-old Caucasian female presented with a slow-growing, solitary bulla on her left upper arm. The bulla first presented one year ago as a pin-sized tan papule. The papule enlarged slowly over one year to its current size of 1.5 cm x 1 cm and now a fluid-filled bulla with a firm nodular core. The bulla was not painful but mildly tender when manipulated. The patient was otherwise healthy and without any personal or family history of similar lesions.

On exam, there was a solitary, transluminating, pink, 1.5cm x 1cm fluid-filled bulla with a solid, nodular core on the left lateral upper arm. [Figure 1] There's a palpable dermal component of the lesion. The overlying skin was normal without telangiectasia, scale, induration, or inflammation. The surrounding skin was characterized by moderate keratosis pilaris. There was no known trauma to the site of lesion. An excisional biopsy of the lesion was performed and gross examination of the specimen revealed a flaccid bulla filled with serous fluid and a white, firm, calcified core. [Figure 2]

What is the diagnosis?

- A. Bullous morphea
- B. Secondary anetoderma
- C. Bullous pilomatricoma
- D. Lymphangioma

Diagnosis

- C. Bullous pilomatricoma

The lesion was excised with a narrow margin of 1mm given its benign clinical behavior: slow growth, no ulceration, and absence of associated systemic findings. Histopathology of the specimen revealed abundant clusters of eosinophilic anucleate "ghost cells" as well as large islands of basoloid cells [Figure 3 & 4] Interestingly, some of the basaloid islands demonstrated a high number (8-10) of mitotic figures per high power field, but given the overall well-circumscribed architecture and the lack of other concerning features, the final pathology was consistent with a bullous pilomatricoma, a rare variant

of a common cutaneous nodule. The excisional surgery was uncomplicated and the patient healed well over the following weeks.

## Discussion

While the other listed choices may present as a blister or bulla, the nodular core is a unique feature of bullous pilomatricoma. Pilomatricoma is a common type of benign adnexal tumor that arises from hair matrix cells and are associated with *CTNNB1* mutation, a gene that codes for  $\beta$ -catenin.<sup>1-3</sup> Bullous pilomatricoma, also referred to as anetodermic pilomatricoma or lymphangiectatic pilomatricoma, is an uncommon variant of pilomatricoma. In a recent study by Li, *et al*, bullous pilomatricoma is found to share molecular features with classic pilomatricoma as a result of the *CTNNB1* mutation.<sup>2</sup> Although no clinical syndromes have been linked to bullous pilomatricoma, traditional pilomatricoma has been associated with Rubinstein-Taybi syndrome, myotonic dystrophy, Gardner's syndrome, and Turner's syndrome.<sup>4-8</sup> Given the clinical, histological, and genetic resemblance between bullous pilomatricoma and traditional pilomatricoma, it is reasonable to bear in mind these syndromic associations when evaluating patients with bullous pilomatricoma.

Histologically, lymphatic congestion, dilation, and resultant lymphedema have been highlighted in the majority of reported cases. Other characteristic histology findings of bullous pilomatricoma include well-differentiated eosinophilic anucleate shadow cells ("ghost cells") and clusters of basoloid cells, both of which were seen in our patient's specimen.<sup>2</sup> Among reported cases of bullous pilomatricoma, the most common site of lesion is the upper arm.<sup>9-10</sup> Based on a literature review by Chen, *et al*, bullous pilomatricoma favors a female predominance and patients between ages of 10-20 years.<sup>10</sup> The goal of this case report is to encourage providers to include bullous pilomatricoma in the differential diagnosis for benign nodular lesions, and it's worth noting that our patient had a classic presentation for bullous pilomatricoma based on current understanding of the condition.

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Fig. 1 Pink, 1.5cm x 1cm, fluid-filled bulla on the left lateral upper arm with background keratosis pilaris.

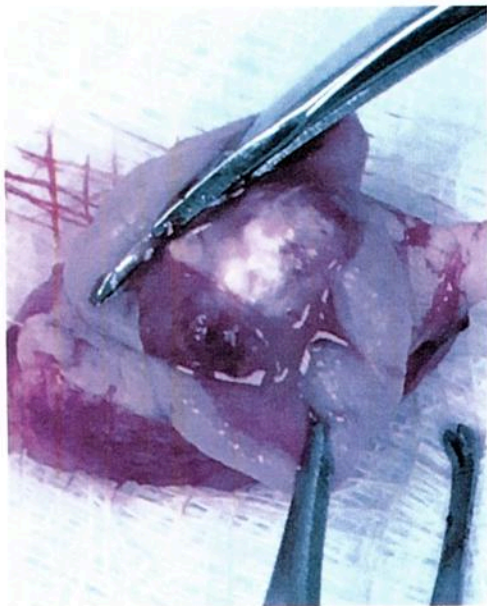


Fig. 2 Gross exam reveals a flaccid bulla filled with serous fluid and a white, firm, calcified core.

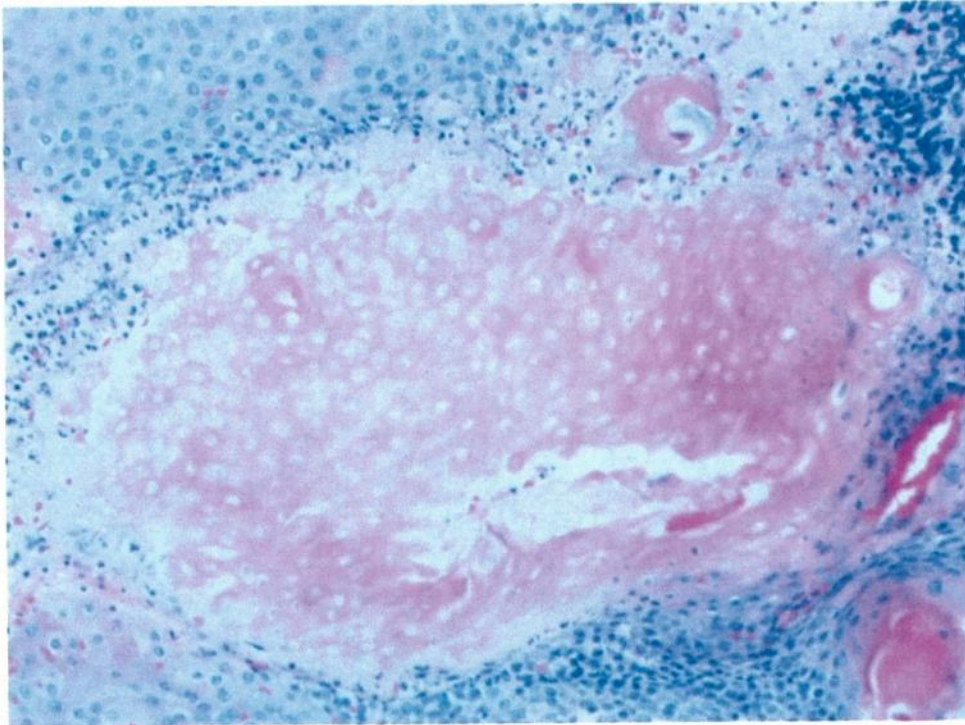


Fig. 3 H&E stain, 20x. Eosinophilic, anucleated shadow cells.

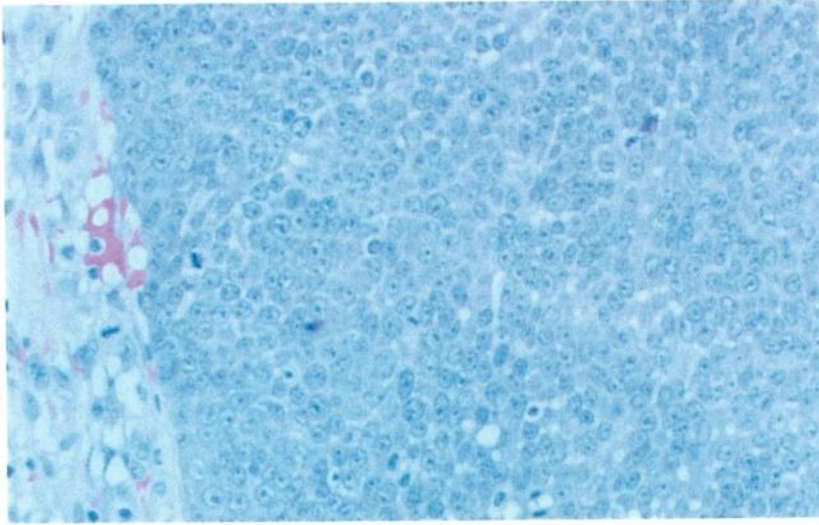


Fig. 4 H&E stain, 40x. Island of basaloid cells.

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